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Partial Nephrectomy in a Cystic Partially Differentiated Nephroblastoma

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Cystic partially differentiated nephroblastoma (CPDN) is a rare neoplastic disorder consisting of a well-demarcated cystic lesion of the kidney where blastemal or other embryonic cells are present in the septa of the cysts. Magnetic resonance imaging can detect the cystic character of the lesion and will produce imaging features that are highly suggestive of either CPDN or cystic nephroma (CN) (synonym: multilocular cyst of the kidney), a benign entity. Although malignant potential exists in CPDN, all cases reported to date have had a favorable prognosis after surgery alone. Partial nephrectomy is considered safe, and the treatment of choice in the newborn period. We report a case of CPDN in a newborn that was successfully treated with partial nephrectomy. More than five years after nephron sparing surgery, the involved kidney shows normal anatomical structure except for a diminished upper pole, no evidence of tumor recurrence and good renal function. Med. Pediatr. Oncol. 28:416–419, 1997.

Key words: cystic partially differentiated nephroblastoma; cystic renal tumors; newborn; infant

INTRODUCTION

In newborns and very young infants abdominal masses are frequently of renal origin. Large series have demonstrated an incidence of 12–20% [1,2,3,4,5,6]. *Clinically the most frequent symptoms are painless abdominal mass, abdominal or flank pain and hematuria.* Tumors of the kidney in this age group differ in several aspects from those of elder children; histology and especially prognosis is in most cases favorable [7]. The well-known relative benign nature of tumors in the first postnatal months should be considered to avoid potentially unnecessary treatment.

CASE REPORT

Ch. H., the second child of healthy parents attracted attention at a routine neonatal physical examination. Otherwise well, he had a palpable mass in the left upper abdominal quadrant. All laboratory examinations, including renal function, were normal and a vannillylmandelic acid screening test was negative. Ultrasound scanning showed a 3 cm spherical tumor of the upper pole of the left kidney with one central and multiple peripheral sonolucent areas separated by hyperechogenic septa (Fig. 1). The renal pelvis was not distended even though parts of neoplasm herniated into the pelvic region. The surrounding renal parenchyma and the contralateral kidney appeared to be normal. MRI very clearly confirmed the presence of a well-defined multicystic lesion (Figs. 2 and 3) with distinctive septa between the fluid filled spaces. The image at the level of renal blood vessels showed a flow void sign within the lumina indicating persistent blood flow. Since all findings were highly suggestive of a benign process, including imaging with Ultrasound and MRI, partial nephrectomy was performed. Intraoperatively the upper pole of the left kidney was found to be occupied by a multicystic mass with a smooth external surface and a well-demarcated boundary between the lesion and the surrounding normal tissue. The involved part of the kidney could be removed completely, and the defect in the collecting system was closed with a running suture. For hemostasis, the resection surface of the renal parenchyma was sealed with fibrin in glue, approximated by a running suture of the renal capsule. The gross specimen measured 3 × 3 × 2 cm and was made up of innumerable cysts (Fig. 4). Microscopic examination demonstrated cystic spaces lined by varying epithelial and cuboidal cells and few highly immature tubular elements and metanephric structures (Fig. 5). As suggested by the earlier imaging, the diagnosis of CPDN was confirmed. Convalescence was uneventful, the preserved part of the kidney was normally perfused and urinary excretion unrestricted. In an over 5 years follow-up, the child has shown normal development without any signs of relapse.

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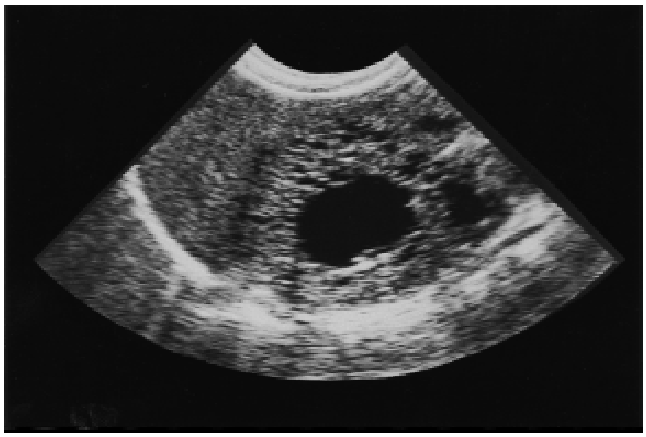


Fig. 1. Ultrasound of the left kidney showing multiple septated sonolucent areas.

DISCUSSION

Edmunds described a first case of multilocular cyst of the kidney in 1892 [8]. Powell and co-workers established criteria for the diagnosis of multilocular cyst of the kidney as solitary lesion composed of a sharply demarcated, multilocular, epithelial-lined cyst which compresses the adjacent renal parenchyma and pelvis without communicating with the latter [9]. In 1956 Boggs and Kimmelstiel modified the criteria: they suggested to exclude all cases with fully developed nephrons within the septa of loculi to emphasize the neoplastic origin of the lesion [10]; a hypothesis which is confirmed by lacking of associated malformation in reported cases and other more recent publications [11,12,13]. In 1974 Brown added the term cystic partially differentiated nephroblastoma (CPDN) in order to describe a lesion which resembled a multilocular cyst of the kidney but with undifferentiated blastema [14]. In 1977 Joshi at al. de-

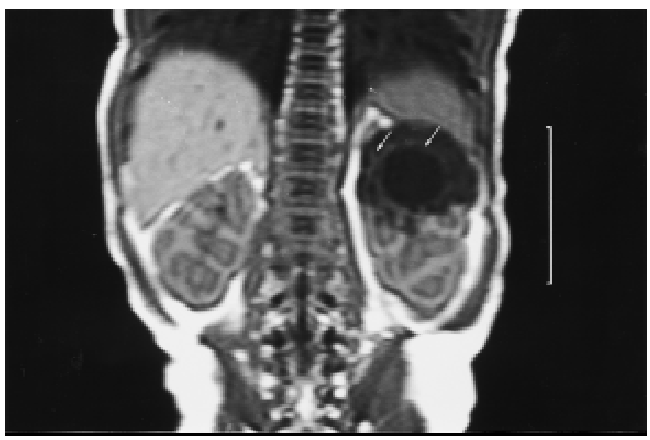


Fig. 2. Coronal T1-weighted magnetic resonance imaging of the upper abdomen. Enlargement of the upper pole of the left kidney by a multicystic tumor. Contents of the cysts with low signal. Septae (arrows).

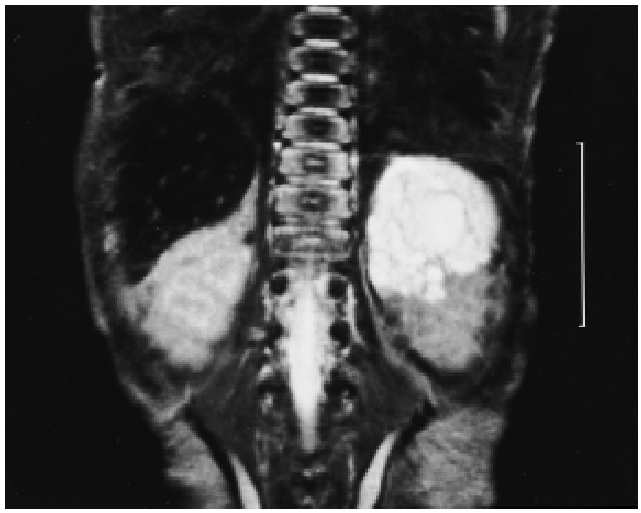
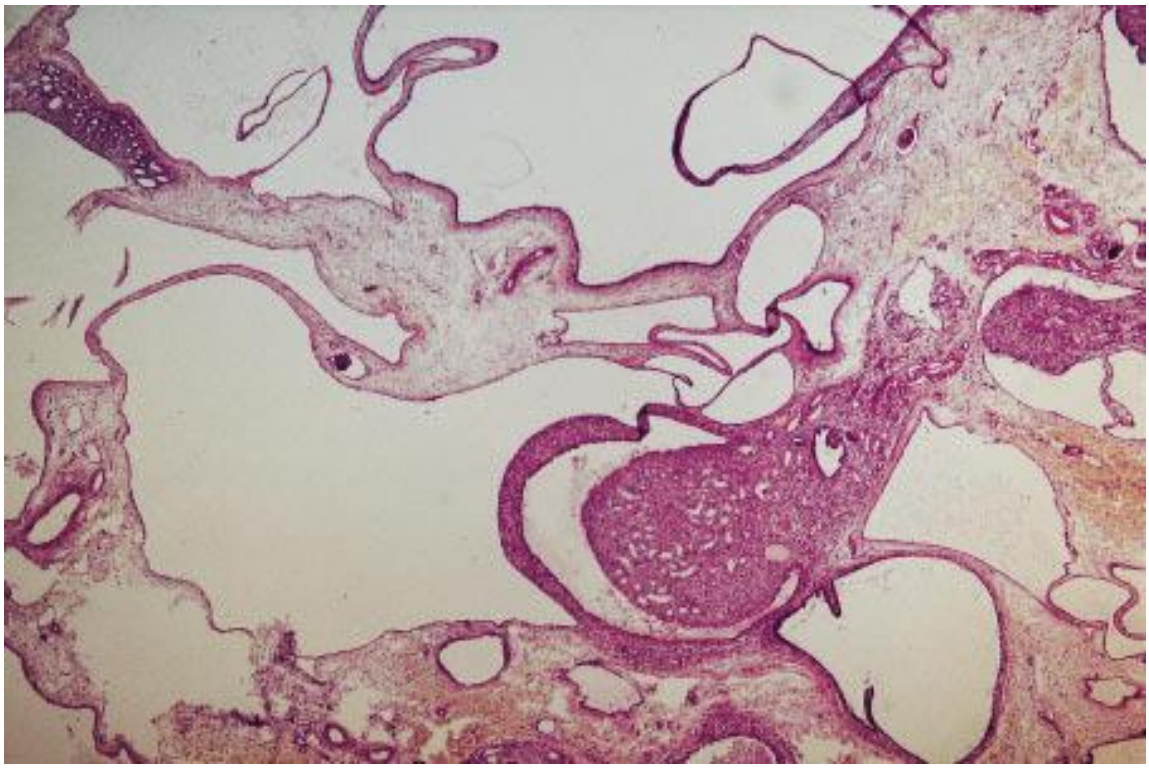


Fig. 3. Coronal T2-weighted image: Contents of the cysts hyperintense and homogenous indicating fluid filled cysts. Septae separating the cystic compartments.



scribed three more cases, with variably differentiated nephroblastomatous elements, which they also termed cystic partially differentiated nephroblastoma [15]. Subsequently a host of terms were used to describe the same entity. Castillo and co-workers listed these multiple and overlapping terms and pointed out the consecutive confusion regarding the clinical importance in their review of literature from 1991 [16]. Today a clinicopathological orientated classification is available, based on the work of Beckwith, Harms, and others [17–22]. In the current opinion multicystic renal neoplasms in newborns and infants form a spectrum of histogenetically related lesions. They are represented by cystic nephroma (CN) (synonym: multilocular cyst of the kidney. *The term cystic nephroma is preferred to point out the neoplastic nature of this entity*) and cystic partially differentiated nephroblastoma (CPDN) [23]. The distinctive histopathological features of these lesions are that they form single renal masses, well demarcated from the unaffected parenchyma and are composed entirely of cysts of varying size that do not communicate with each other or the renal pelvis. According to the classification scheme of Joshi and Beckwith, CPDN contains immature cellular elements in contrast to CN, as demonstrated in our specimen. Harms and coworkers have added a further entity named cystic nephroblastoma characterized by abundant blastema and undifferentiated tubules and summarize all three entities into the group of ''low-risk'' nephroblastomas. Following Beckwith, CN forms the benign end of the tumor spectrum, CPDN being an intermediate category, while polycystic Wilms' tumor forms the malignant spectrum. To prevent inadequate therapeutic approaches a preoperative detection of a solid component in cystic renal tumors is of high importance: cystic changes may occur in otherwise solid masses due to tumor regression or as result of cystic dilatation of tubular structures. Ultrasonography is an easily available method with a high accuracy for cystic lesions and thus the screening method of choice [24]. However, MRI better reflects the characteristic morphology of the tumor and thus can strongly suggest the preoperative diagnosis [25–28]. Hemorrhage, possibly mistaken as an area of solid tumor in ultrasound, can be differentiated on MRI by means of the higher signal intensity on T1 weighted images.

Large series have demonstrated that cystic renal tumors in neonates and young infants are to be considered ''low-risk'' nephroblastomas. Chemotherapy and/or radiation in neonates is associated with a higher incidence of immediate complications, especially infections. It can also cause damage to the immature tissue and can lead to major impairment of the survivors [29]. A dilemma occurs in patients where relatively undifferentiated cellular elements are found within the cyst. To clarify whether these patients benefit from adjuvant therapy or not, Castillo et al., Schmidt et al., Joshi et al. and Beckwith et al. reviewed their own cases and the literature. Their results, together with other newly published case reports demon-

strate that the vast majority of cases of CN, CPDN and cystic nephroblastoma show no evidence of tumor recurrence or metastasis despite the fact that no chemotherapy and radiotherapy had been administered [30–35].

Due to the benign clinical course of these tumors during the first months of life the therapy of choice is nephron sparing surgery without further treatment. The tumor resection should be complete, avoiding rupture of the neoplasm and with a security margin of at least 1cm. Armed with modern diagnostic tools like ultrasonography and MRI, as well as an understanding of nature of renal cystic lesions in the newborn and very young infants, an accurate preoperative diagnosis is achievable.

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